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Massive Intra-atrial Wilms' Tumor: A Treatment Dilemma

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Key words: Wilms' tumor; CT scan; hepatomegaly; intra-atrial

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The patient is a 5-year-old boy who presented with vomiting, abdominal pain, and distention. On physical examination, the patient was in moderate respiratory distress and was found to have bilaterally decreased breath sounds, abdominal distention, a leftsided abdominal mass, and hepatomegaly.

Tim Brown, MD (Pediatric Radiologist), R. Morrison Hurley, MD (Pediatric Nephrologist)

An abdominal CT scan revealed a leftsided abdominal mass arising from the left kidney and extending into the left renal vein and inferior vena cava (Fig. 1). It also showed hepatic venous congestion with hepatomegaly and ascites. The chest CT showed right atrial extension of the mass with large bilateral pleural effusions. No parenchymal metastases were seen. A percutaneous needle biopsy of the left kidney was attempted with no success in obtaining tumor tissue.

Marc D. Le Gras, MD, Herschel C. Rosenberg, MD (Pediatric Cardiologists), Alan H. Menkis, MD (Cardiac Surgeon), Yousif H. Matloub, MD (Pediatric Oncologist), A. Rashid Dar, MD (Radiation Oncologist).

A two-dimensional echocardiogram was performed and showed a massive intra-atrial tumor that measured 1.7 cm $\times$ 1.9 cm and occupied $1/3$ to $1/2$ of the right atrial cavity (Fig. 2). The patient was transferred to the intensive care unit, placed on oxygen therapy, and bilateral chest tubes were inserted. Surgery was initially contemplated, but was deemed too hazardous, so the patient was immediately started on chemotherapy as per the National Wilm's Tumor Study IV protocol, regimen DD-4A. Accordingly, he received dactinomycin (45 mcg/kg) and vincristine (1.5 mg/kg).

A follow-up echocardiogram performed 3 days after starting chemotherapy showed the mass to have roughly doubled in size. It was now seen to be occupying most of the right atrial cavity and crossing the tricuspid annulus (Fig. 3). Radiation therapy was then initiated with the hope that in combination with chemotherapy, it would halt the tumor's rapid growth. A total of 1,000 cGy was given to the mediastinum with lung shields and paraaortic region down to the sacroiliac area, with shielding to the right kidney (Fig. 4). Radiation therapy was delivered in 10 fractions with a 4 mv photon unit.

Five days after starting chemotherapy, the patient developed seizures and a transient encephalopathy. These were thought to have been caused by vincristine in a background of worsening liver functions, as the patient's hepatic transaminases and serum bilirubin were found to have coincidentally increased from their pretreatment values. The seizures were readily controlled with phenytoin, and the encephalopathy resolved in the following few days.

Chemotherapy was continued for 10 more weeks. During that time, the patient improved steadily and was transferred to a regular ward. The atrial extension and cardiac function were regularly followed by two-dimensional echocardiograms. The atrial mass decreased gradually in

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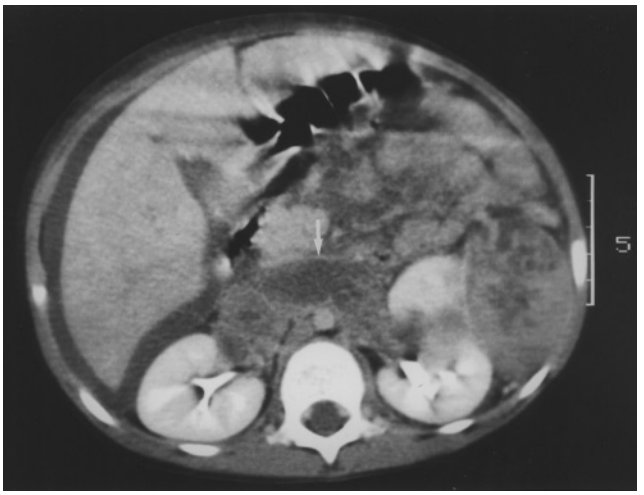

Fig. 1. CT image at the level of the left renal vein at the time of diagnosis shows tumor extending across the renal vein to the inferior vena cava (arrow). Note large tumor extension lateral to the kidney. Ascites is seen around the right lobe of the liver.

size, reaching preoperative dimensions of 1.6 cm × 1.4 cm from 3.3 cm × 2.6 cm prior to radiation therapy (Fig. 5). The abdominal portion of the tumor, however, underwent a relatively greater decrease in size (Fig. 6).

Alan H. Menkis, MD (Cardiac Surgeon), David Girvan, MD (Pediatric Surgeon)

A thoraco-abdominal approach was used, and the child underwent left nephrectomy successfully. Attention was then directed to the right atrium and inferior vena cava. Right heart bypass was utilized as opposed to full cardiopulmonary bypass in order to avoid the possibility of disseminating tumor cells into the systemic circulation. Also, in addition to the gas exchange membrane, two arterial line filters were inserted in series into the bypass circuit to enhance filtration of particulate matter. The right atrial tumor was removed by dissecting it from the endocardium to which it was attached. The inferior vena cava had been largely obliterated and subsequently recanalized. A thromboendarterectomy was performed. Biopsies were obtained from the atrial, caval, and renal masses. The atrial and caval samples were composed of organizing thrombus; that from the renal vein was mostly fibrotic. The left renal sample contained an area of favorable histology nephroblastoma and a focal area of nephroblastomatosis.

DISCUSSION

Dr. Matloub. Concomitant treatment with chemotherapy and radiation was required to achieve a life-saving reduction in the size of the intra-atrial extension, thus allowing a safer elective surgery to be performed at a later date. The need for irradiation makes our case exceptional and calls attention to the good results obtained with this modality. Although it is agreed that irradiation in pediatric patients should be kept to a minimum, it should not be omitted when clinical pressures warrant its use.

Intracardiac extension is a rare event in nephroblastoma, occurring in ~0.7% of cases [1]. Although the usual initial management has been to excise the atrial mass along with the vena caval extension [1–6], there have been 18 reported cases treated initially with chemotherapy [7–9]. In all these cases, however, no mention was made of the size of the atrial mass nor of any hemodynamic compromise that it may have caused. Chemotherapy alone is often successful for this purpose, but that was not the case in our patient. Rather than shrinking or at least stabilizing in size, the tumor grew during chemotherapy. We were presented with a massive intra-atrial extension of a nephroblastoma in a 5-year-old boy that doubled in size in 3 days and came to occupy most of the right atrium and crossed the tricuspid annulus. Surgical intervention was deemed to be too hazardous in this child. Therefore, our only option was medical management, which had to act promptly in view of the patient's critical condition.

Addendum

Dr. Matloub. The patient completed the 24 weeks of chemotherapy as per the above mentioned protocol and is currently disease-free and doing well 4 months from end of three-drug chemotherapy, i.e., dactinomycin, doxurubicin, and vincristine.

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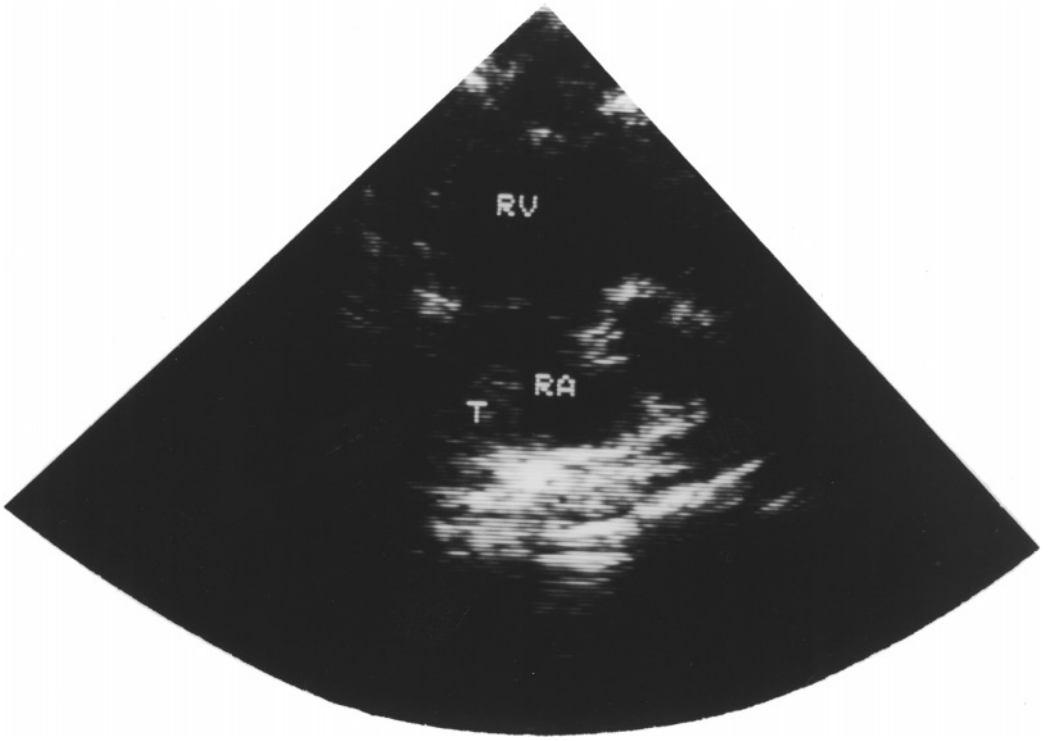

Fig. 2. Two-dimensional echocardiogram performed at diagnosis shows tumor (T) in right atrium.

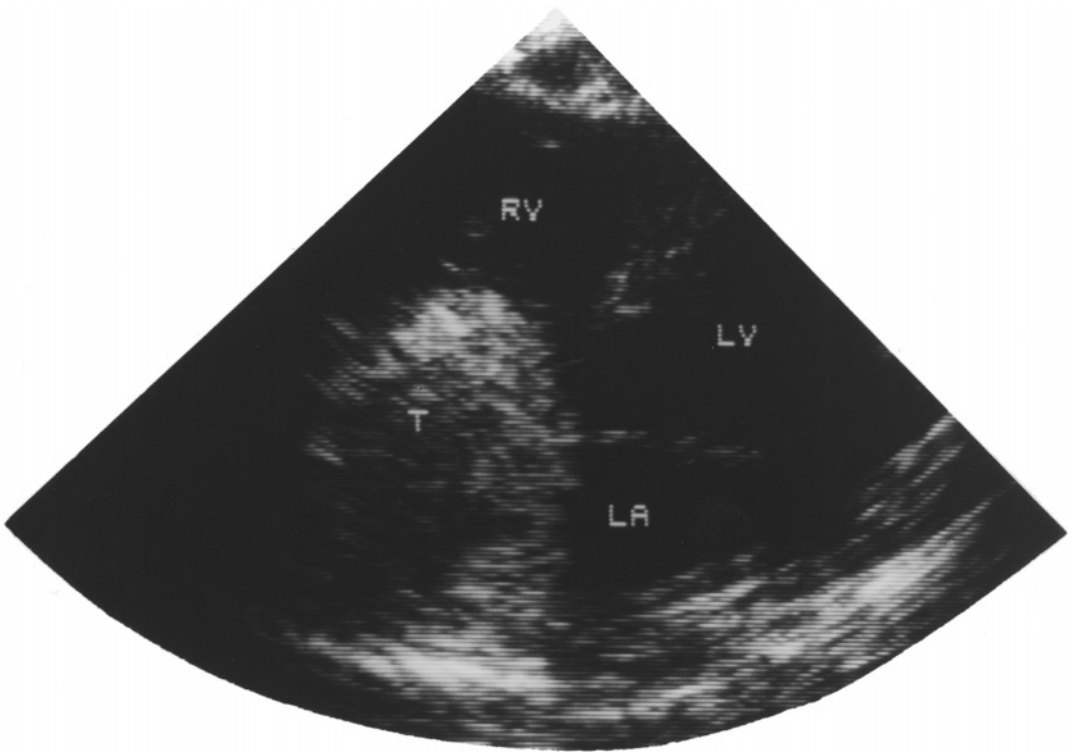

Fig. 3. Two-dimensional echocardiogram performed 3 days after starting chemotherapy shows the intraatrial mass occupying most of the right atrial cavity and crossing the tricuspid annulus.

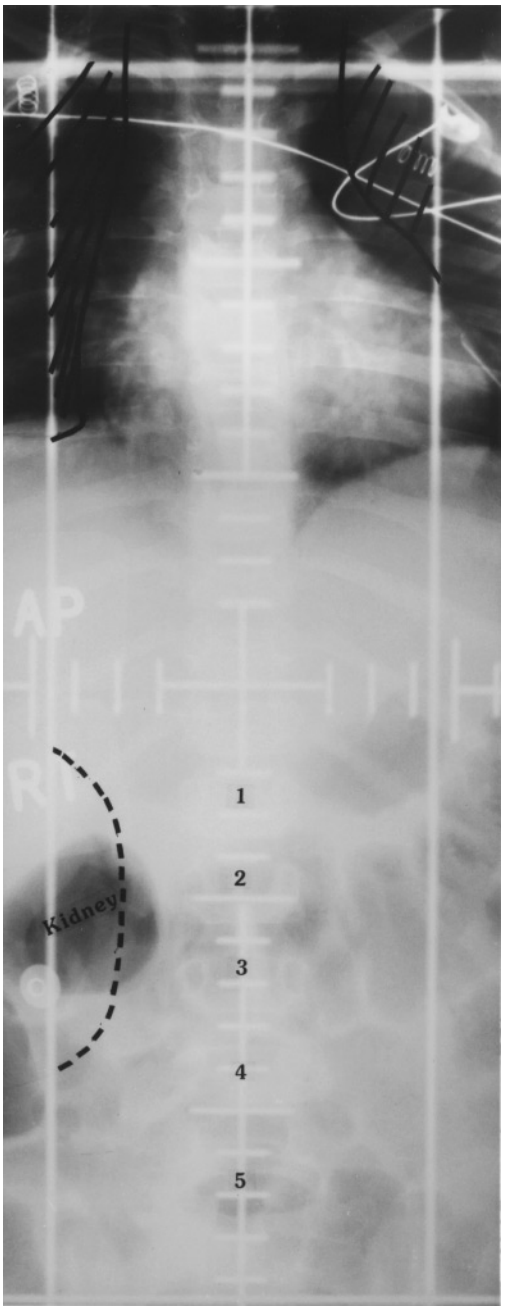

Fig. 4. Radiation field included the mediatinum, paraortic region extending to the sacroiliac area, with shielding to the lungs and right kidney.

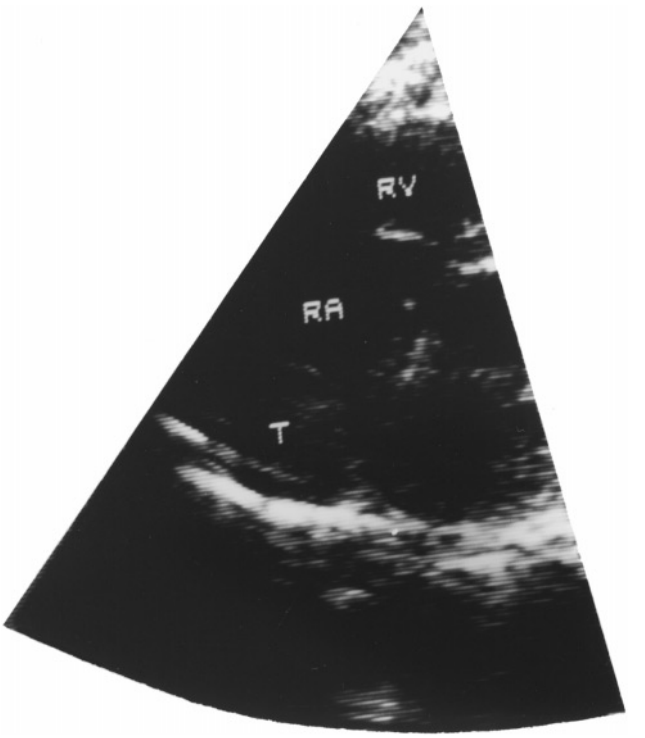

Fig. 5. Two-dimensional echocardiogram performed prior to surgery showing intraatrial tumor shrinkage in response to combined therapy.

Fig. 6. CT scan at the level of the renal vein (arrow) performed prior to surgery showing response to treatment.

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Series Editor's Note

The Tumor Board Proceedings in this issue comes from London, Ontario.

Many of the names of English, and therefore North American, cities recall the nearly three centuries of the

Roman presence in Britain starting in A.D. 43 and their Latin names during that period. Thus, London was *Londinium* (from the Celtic), which became one of the major Roman centers. Lincoln was *Lindum*, a *colonia*, or retirement settlement for legionnaires, many of whom remained on the island after their tour of duty ended. The Latin *castra* for camp is echoed in the *-chester* or *-cester* ending of many other English place names, e.g., Gloucester, Leicester, Colchester, as well as Chester itself. A small but excellent museum depicting Roman life in Britain is to be found in *Cirencester*, once *Corinium*, now a small English city, but then second only to London in importance. All of which calls to mind Winston Churchill's quip (from the Latin *quippe* = indeed) concerning the Roman occupation. In paraphrase, he wrote: "Never have Britons been so well governed and never have they been so unhappy," a sentiment he contradicts in his *A History of the English Speaking Peoples*, Vol. I, Dodd, Mead, New York, 1956. There he says (p. 35), "For nearly three hundred years Britain . . . enjoyed in many respects the happiest, most comfortable, and most enlightened times its inhabitants have ever had." The truth probably lies somewhere in between, when one recalls the frequent insurrections by the indigenous tribes notably that led by Queen Boadicea in A.D. 61.

The importance of a pronunciation (Latin = *pronuntiatio*) mark is shown in "Canada." That great and beautiful country is reduced to a ravine in Spanish by the addition of a circumflex (*circum* = around + *flectere* = to bend); thus canada = cane, a place where canes grow. Ravine is from the Latin: *rapere* = rapine, to rob or steal, here used to mean "to wear away" since a ravine is a narrow and deep cleft eroded over time by the running waters of a stream.